

CYSTIC DEGENERATION OF THE KIDNEY.

BY CLARENCE M. NICHOLSON, M.D.,

OF ST. LOUIS, MO.

Professor of Practice of Surgery and Clinical Surgery, Medical Department,
St. Louis University.

CASE.—Mrs. H. M. J., age 46, married, referred to me by Dr. W. C. Lewis, was admitted to the Rebekah Hospital August 13, 1906. During the five years previous she had complained of difficulty in breathing which at times was very severe, the attack would last from one half to four hours. Whiskey and other home remedies were made use of and on some occasions artificial respiration was resorted to. She occasionally complained of nausea, but never vomited. Five months ago she noticed an enlargement in the left lumbar region which rapidly increased in size.

Examination.—Urine from left kidney highly colored, specific gravity 1028, sediment, normal, blood, slight in amount, casts, both hyaline and granular. Urine from right kidney normal. A mass extending from the symphysis pubis upward to the left, behind the lower ribs, could be made out; approximately six or seven inches in diameter and twenty-one inches in length. There was some tenderness.

Operation.—A median incision showed the opposite kidney was not enlarged. A right lumbar incision exposed a multilocular cystic kidney; the visible cysts varying in size from a split pea to an orange. The size of the mass precluded the possibility of delivery through the incision in the loin until after thirty or forty cysts had been punctured. The vessels and ureter were separately ligated, the remaining mass removed and the cavity packed with sterile gauze. Convalescence was uneventful, the patient leaving the hospital six weeks later; after eighteen months she remains entirely well.

Under the name Cystic Degeneration, has been described a peculiar lesion manifested by the presence of multilocular cysts, occupying the area of and replacing the kidney. Frequently cystic kidneys contain no discernable renal tissue; in

other instances only part of the kidney is involved. The disease often exists for years; no diagnosis being made owing to entire absence of symptoms, while not infrequently sudden death takes place from uremia or cerebral hemorrhage due to kidney insufficiency.

Cystic kidney in the new-born has long been recognized. Fussell collected eleven cases in which it was necessary to mutilate the fetus in order to accomplish delivery. The tumors in the adult may be of enormous bulk weighing (as in Hare's case) as much as twenty-four pounds. These tumors are usually bilateral and may increase in size under observation.

Hematuria, sclerosis of arteries, hypertrophy of heart with accentuated second sound, and albuminous urine are among the more common symptoms. Cystic kidney occasionally, as in the case here reported occupies a large portion of the abdominal cavity. It is made up of numerous cysts varying in size from a pin head to a cocoanut and little or no recognizable renal tissue may be found. The fluid within the cysts is clear, slightly albuminous, presenting cholesterol, blood pigment and detritus resulting from degenerative and necrotic processes in the epithelium of the cystic wall. In some instances the connective tissue of the cyst is not covered by a recognizable amount of epithelium, in other instances epithelium, when found, varies in amount, both granular and necrotic; and in still other specimens columnar epithelial cells from the inner layer of the cyst wall are found. The matrix between the epithelial cell may be fibrous or myxomatous and is not infrequently extremely vascular.

Our lack of knowledge of the etiology of these cysts has given rise to much discussion. The earliest theory, that of Virchow, was that they were due to an obstruction of the tubules. Later he expressed a belief that cystic kidney was due to an intra-uterine papillitis, this view was reaffirmed by him in 1892.

Arnold, regarded the process as beginning in the pelvis of the kidney and speaks of the lesion as an "ascending pyelopapillitis fibrosa." The inflammatory origin of cystic

kidney, founded on the great increase of connective tissue in the pyramids and the numerous foci of round-celled proliferation present has been supported by Thorne.

Brigidi and Severi, in 1880, basing their opinion upon epithelial sprouts into the surrounding connective tissue from the walls of the tubules, especially of the straight tubules, also an increase in the layers of the tubules and proliferations of epithelium in continuity, so that the epithelium appears contorted within the lumen, claimed that cystic kidney bore marks of a tumor and called it multilocular adenocystoma, this point of view though confirmed by Chotinsky, Nauwerck, Hufschmidt, Janowski, Kalilden, Hain and Alber was declared by Marchand to be untenable. Durlach believes it due to a proliferation between the lobules of the kidney, that is, of tubules between the pyramids and their outlying cortex. Leichtenstein thinks it begins as an inflammation or inflammatory irritation in the arteriolæ rectæ, which are small vessels between straight tubules and give striations to pyramids.

Shaddock concludes that the condition depends on the mal-development of mesonephron or the Wolffian body fused with the metanephron and that the cysts result from the evolutionary changes in the included mesonephron. Van Mutach recognized from a microscopic point of view striking embryonal characteristics of cystic kidney.

Hildebrandt explained the condition on purely embryological grounds. Deetmar, Schenkl and Rukert accept cystic kidney simply as a form of mal-development of the organ.

Milward divides the clinical history of the disease into three stages:

1. The stage of progressive enlargement of one or both kidneys without subjective symptoms. The renal enlargement is discovered, if at all, by accident. This stage may last from a few months to several years.

2. The stage of subjective symptoms and objective signs. This stage lasts from a few months to seven or eight years, or even longer. The signs and symptoms are dependent on the size and weight of the tumors.

3. The stage of decreasing elimination of urine. In this stage appear the symptoms of uremia, or cerebral complications. This disease which affects males and females in about equal proportion usually terminates fatally about the age of forty-five. The symptoms of the second stage are dull, aching pain in the region of the kidney. Flatulence, headache, dyspepsia, vomiting, anorexia and constipation are frequently met with. Examination of the urine may throw very little light on the diagnosis, though the specific gravity is usually low and the amount passed is generally slightly increased. A trace of albumen is usually found, but is never present in large quantity in the second stage except when there is a large amount of blood. Leucocytes may be met with in large numbers and pus is often present. Blood is usually present and may occur at intervals of weeks, months or years or may never be noticeable by macroscopical examination of the urine, though according to Milward it is always present and is significant. The symptoms of the third stage are those of uremia.

The treatment for congenital cystic kidney consists of removal of the organ, unless the other kidney is diseased which can only be positively determined by exploratory incision. While Schmidt, Neimeyer, Bardeleben, Hayems and others have performed nephrectomy for polycystic kidneys, the patients having died of uremia, the opposite kidney has in all cases been found to be the seat of rather extensive disease. Where the opposite kidney is involved an incision through the loin, puncturing the cysts and stitching the kidney to the lumbar muscles is the procedure indicated.

Pathological Examination of the Specimen Removed in the Case Reported.—GROSS. The kidney after evacuation of the larger cysts weighed 1,400 gms. It is 24 cm. in length and 15 cm. in width, and the shape is almost rectangular. The anterior surface is studded with small cysts varying in size from almost microscopical points to the size of a walnut. (Fig. 1.) They bulge outward prominently, giving the kidney the appearance of a bunch of grapes. The superficial cysts have very thin walls and are transparent. They are light yellow to brownish-yellow in color. The posterior surface is similar to the anterior; however, the cysts are not so prominent. The pelvis of the kidney is narrow and extends deep between

the upper and lower overhanging projections of cyst masses. The vessels entering are large, but not thickened. The ureter is not dilated.

In making the incision for the cut surface, the organ is laid open in its longest diameter, the incision being made from the convex surface toward what constitutes the pelvis. This cut passes thirty-three cysts, each larger than a pea. (Fig. 2.) The cut surface of the kidney shows little kidney tissue recognizable as such. No striations can be seen. The surface has a honeycombed appearance. The color of the kidney substance is grayish. The largest cyst is at the upper, inner pole, and has a diameter of $7\frac{1}{2}$ cm. It is deep, round, and lined with a smooth surface, and crossed by falciform trabeculae in part. It has five depressions, about $1\frac{1}{2}$ cm. in diameter, which have the appearance as if five small cysts had coalesced and formed the large one. Shining through the wall of the lower inner depression are seen two black cysts. The contents of the cysts are in part clear serous fluid; others contain a jelly-like colloid material. Some contain a purulent material, and others a black hemorrhagic fluid, or are clouded with a brown substance. Numerous smaller cysts are seen on the cut surface of the kidney. They are mostly round and form hollow spaces. Their septa and walls show traces of kidney parenchyma. The cyst contents is alkaline in reaction and contains albumin. Microscopically it shows red blood cells and leucocytes with granular degeneration. Fat is shown to be present by Sudan III stain. The calyces are only rudimentary and are distorted by compression of the cysts. Their lining is smooth and pink in color. The walls are thicker than normal. There is considerable pelvic fat.

MICROSCOPIC: Section 1.—Showing one of the smallest cysts. The cyst content is not stained. The cyst is lined by tubular epithelium which is slightly flattened. The protoplasm of the cells is granular and not stained as deeply as normal. The nuclei show a vesicular condition. They are nearly all large and contain less chromatin than do the nuclei of normal tubular epithelium. The connective tissue surrounding the cyst is increased, and is rich in nuclei. The tubuli contorti are increased in number and apparently widened. Their lining epithelium is wider than that lining the cyst and takes a deeper stain. (Figs. 3 and 4.) The glomeruli do not differ from those of a normal kidney. They are normal in number and size. Their capsule is not thickened.

Section 2.—Showing wall of a microscopic cyst smaller than a pea. (Fig. 5.) The content is not stained. The lining epithelium is much flattened and faintly stained. In places it is detached or absent. The connective tissue shows greater increase in nuclei than in previous section and replaces tubules to some extent. The picture is similar to that of a chronic interstitial nephritis. There is marked infiltration of lymphocytes. The tubules are elongated and flattened so that their lumina are filled. The glomeruli are distorted and deformed by pressure and the capsule of Bowman is thickened. Some glomerular tufts show vacuolization. It is probably a hydropic degeneration, however; no fat is seen by staining with Scharlach. The blood vessels are thickened; chiefly the tunica

muscularis. The lumina are large and filled with blood in the region near cysts.

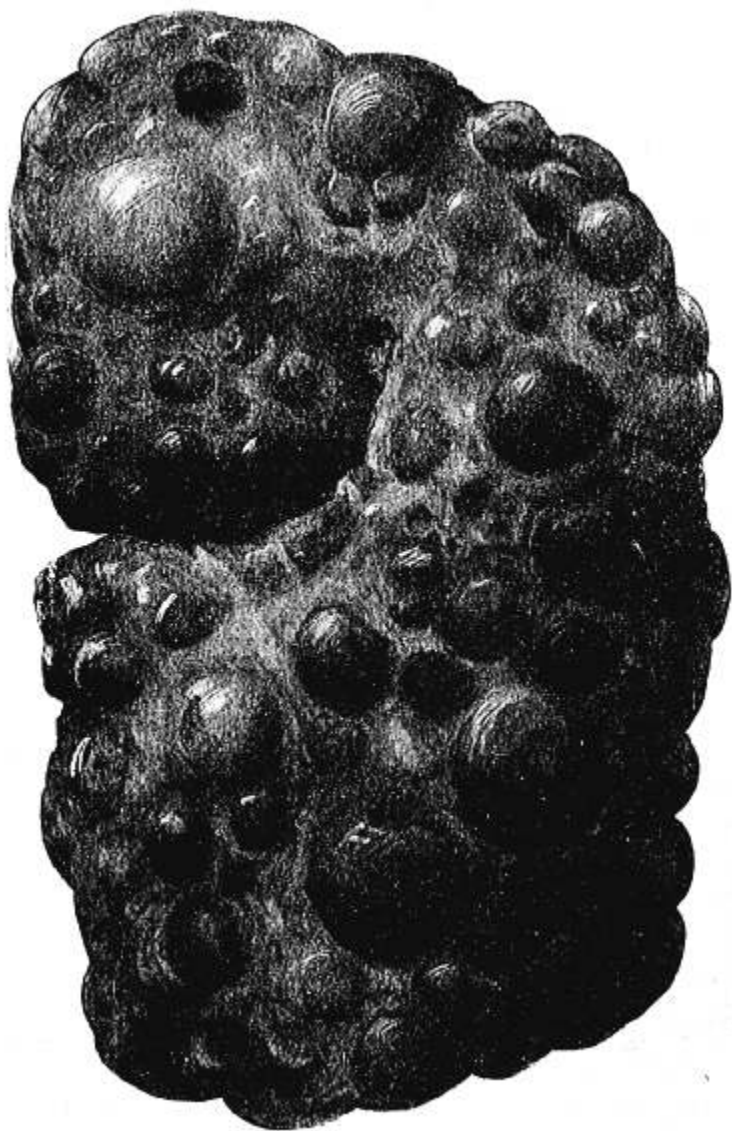
Section 3.—Showing wall of a hemorrhagic cyst the size of a pea. (Fig. 6.) The content consists of blood pigment, degenerated epithelial cells and detritus. The epithelial lining of the cyst is absent, so that only a connective tissue lining is seen, which is wide and shows irregular heaps of new-formed nuclei. There are no tubules present. Some glomeruli are present, but show a more advanced stage of degeneration than in the previously described sections. They are small with greatly thickened capsules. The tufts fill the capsule only in part. The blood vessels are greatly dilated.

CONCLUSIONS.

I. In the cells lining the cysts, the entire row are alike, and apparently in the same stage of secretion, that is, the nuclei are poor in chromatin and show a vesicular condition. In the normal convoluted tubules, this condition is present in a few cells only while very many are rich in chromatin and show filaments or striations known as "ergastoplasm." This condition of the normal convoluted tubules is met with in active secreting cells. While the condition of the cells lining the cyst would correspond to a stage in secreting activity, it is not probable, however, that the entire row lining the cyst would show the same stage in secretion because we have no proof that secretion occurs in cells simultaneously. Therefore in my opinion the cells lining the cysts are less active in secreting power than normal epithelium.

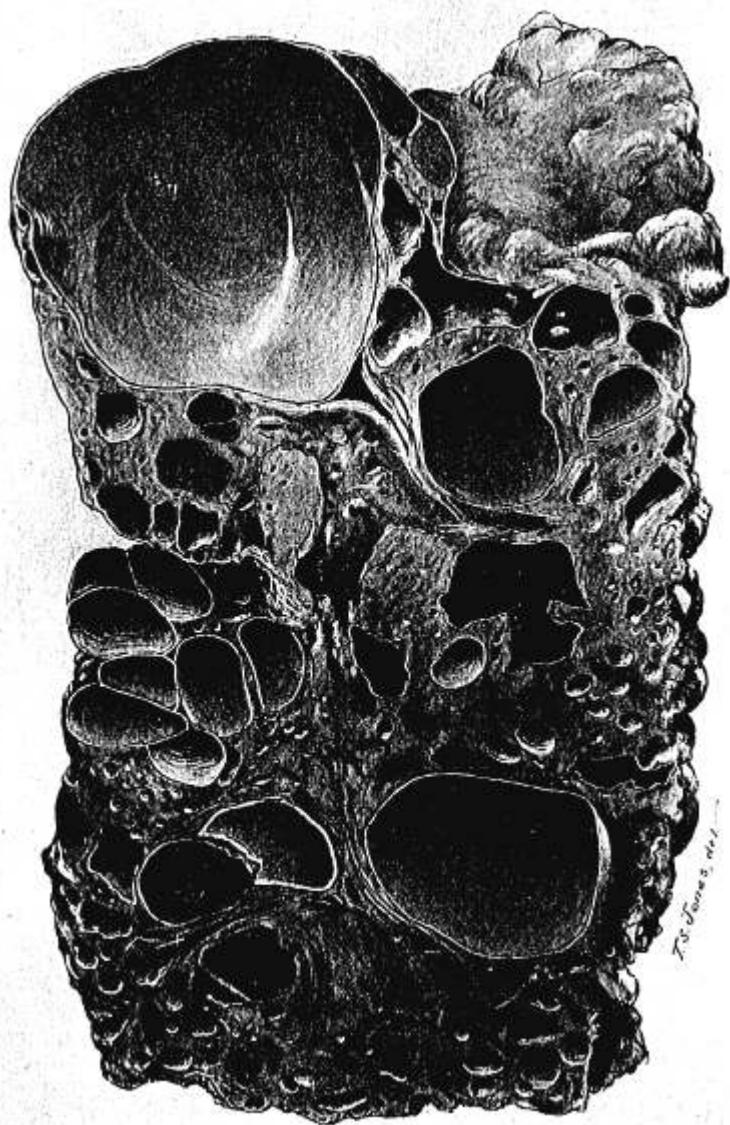
II. These various theories of the origin of cystic kidneys have been discussed previously in this article. From the study of the microscopical sections I find that the smallest cysts are lined with the same epithelium as that lining in the convoluted tubules. Therefore, one must conclude that the cysts originate in the convoluted tubules by dilatation and proliferation, lining epithelium. In this kidney I can say with certainty that the cysts do not arise in the glomeruli, the blood vessels or the connective tissue.

FIG. 1



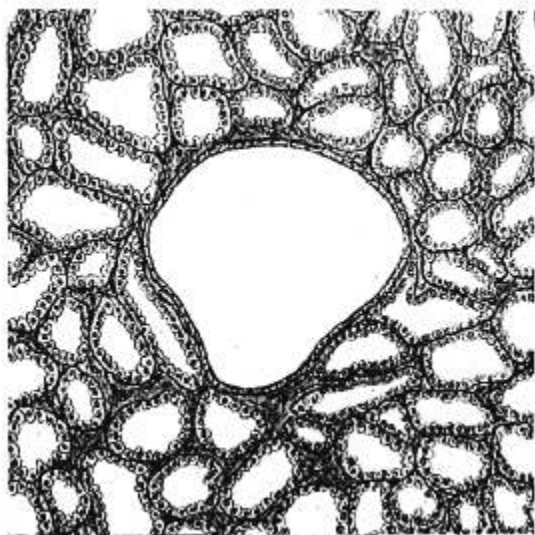
External surface of cystic kidney extending from the brim of the true pelvis to the angle of the scapula. Removed at operation from patient of Dr. E. K. Lewis. Recovery.

FIG. 2.



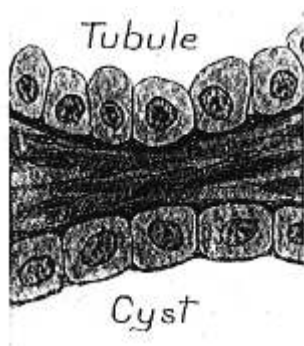
Transverse section of Fig. 1.

FIG. 3.



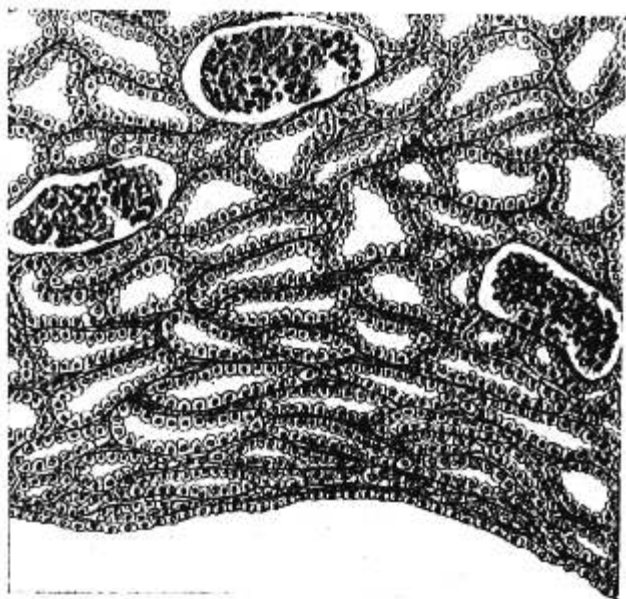
Microscopical appearance obj. 3. of one of the smallest cysts from section 1.

FIG. 4.



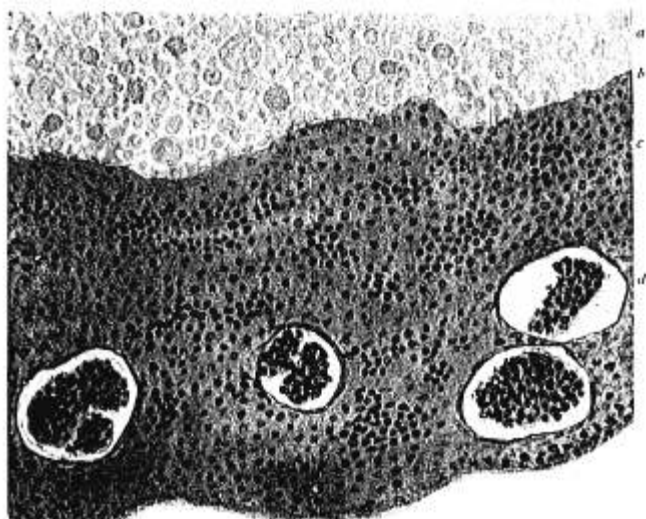
High power obj. $\frac{1}{2}$ showing lining of cyst and tubule, from section 1.

FIG. 5.



Microscopical appearance of wall of a cyst smaller than a pea, from section 2. *a*, cyst lining *b*, flattened tubules.

FIG. 6



Microscopical appearance of wall of a hemorrhagic cyst the size of a pea, from section 3. *a*, blood pigment. *b*, connective tissue lining. *c*, round cell infiltration. *d*, degenerated glomeruli. No convoluted tubules are seen.